



CASE REPORT

Isolated double chambered right ventricle as a rare cause of sudden death in infancy

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KEYWORDS

Sudden death;
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ventricle;
Autopsy**Abstract** Isolated double-chambered right ventricle (DCRV) is a rare form of congenital heart disorder in which the right ventricle is divided into a high-pressure inlet portion and a low-pressure outlet portion by an anomalous muscle bundle. Rarely, autopsy cases describing this congenital heart disease have been reported in the world literature.

We report the case of a 50-day-old male who died seven days after been suffering from fever and dyspnea. The autopsy revealed essentially an isolated double-chambered right ventricle (DCRV) and an important pulmonary edema.

In this paper, we present the first reported case of sudden unanticipated death in infancy due to isolated double-chambered right ventricle.

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1. Introduction

One of the leading causes of prenatal and infant death from congenital malformations is congenital heart disease (CHD).¹ Its incidence in different studies varies from 4/1000 to 50/1000 live births.² A recent report from the United Kingdom Northern Congenital Abnormality Survey showed that 10% of deaths in this pediatric cohort with at least one congenital anomaly were associated with CHD.³

Double-chambered right ventricle is a rare congenital heart disease which consists of a cardiac disease of the right ventricular outflow tract obstruction characterized by an anomalous muscle bundles (AMB) that divides the right ventricle into

two chambers: a high-pressure inflow chamber and a low-pressure outflow chamber.

We report an unusual cause of death in a 50-day-old male infant showing a double-chambered right ventricle in the autopsy.

2. Case report

A male neonate born at 35 weeks after a well attended pregnancy with a normal vaginal delivery to a 25 year old primigravida mother suffering from toxemia of pregnancy. Apgar scores were 8 and 9 at 5 and 10 min, respectively. Birth weight was 2800 g. The initial clinical exam was normal.

On day 43 of life he was suffering from fever and dyspnea. A chest film was required showing no abnormal features. CRP was little high and hemogram test and electrolytes were normal.

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In view of sepsis, antibiotics were started. The condition continued to be critical and the clinical status evolved to the worst.

Although he was transferred to an intensive care unit, the baby deceased on his 50th day of life.

Because there was a dubiety regarding the manner of death, a forensic autopsy was requested by the district attorney almost 17 h after death.

The deceased was a well-developed, well-nourished, 3.664 g, 54 cm male infant without any evidence of trauma.

The external examination revealed a lip and subungual cyanosis, some bloody secretions from nostrils, further there was no visible injury or congenital malformations in the external examination of the body.

On autopsy, the 58 g heart (the expected weight is $22 \text{ g} \pm 10 \text{ g}$) was enlarged and hypertrophied and had smooth epicardial surfaces with a few petechiae. It measured $3.5 \text{ cm} \times 5 \text{ cm} \times 7.5 \text{ cm}$.

The great vessels of the heart had no anomalies or developmental defects. The coronary system was left-dominant, with a normal caliber and without any atherosclerotic lesions. On the opening of the heart, the left ventricular wall was 0.83 cm in thickness and of mottled appearance and the right ventricle was widely hypertrophic; 0.43 cm in thickness (Table 1). We notified the presence of a muscle bundle measured 1.1 cm in large that protrudes from the right ventricular free wall to the interventricular septum (Fig. 1). It divides the right ventricle into two chambers.

No apparent sclerosis of the cardiac valves and no stenoses, thrombi, or anomalies of the coronary arteries were noted. The foramen ovale was closed, and we could not find any other cardiac anomalies.

The 70 g left lung and 85 g right lung had smooth pleural surfaces with multiple scattered petechiae and congested parenchyma.

Bronchi contained thick bronchial secretions and pulmonary arteries were free. The pressure of the pulmonary sections showed an issue of purulent secretions associated with massive edema.

The rest of organs, such as the brain, the liver, and the kidneys, were extremely congested without any other macroscopic abnormality.

A histological examination was performed with hematoxylin and eosin stain.

The endocardium showed pronounced thickening with laminar deposition of elastic and collagen fibers prevailing at the right ventricle. There was no fibrosis or necrosis. The conduction system had no significant lesions. The section of the right ventricular myocardium demonstrated marked myocyte hypertrophy and disorganization. The muscle bundle

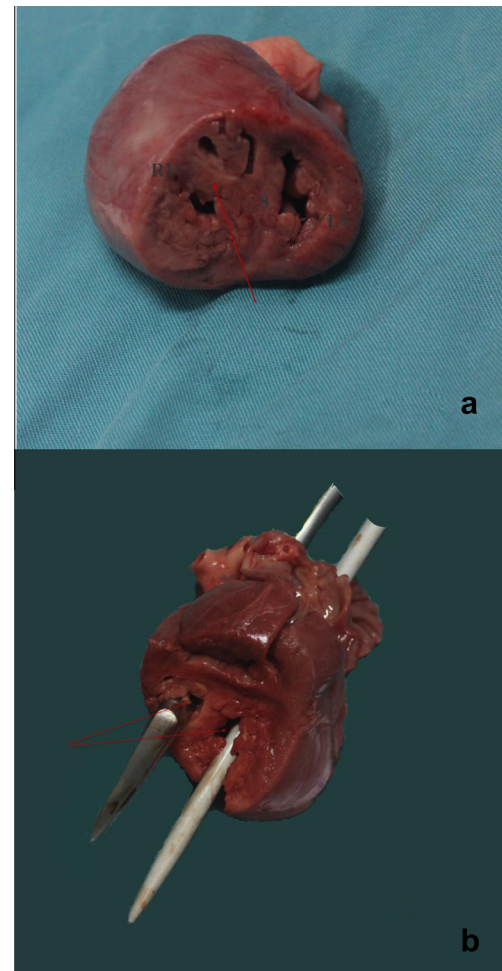


Figure 1 (a) and (b) The right ventricle (RV) is divided into two chambers by a pseudo septum.

dividing the right ventricle was composed by whorled myocardial fascicles. The myocardial cells had hypertrophic nuclei (Fig. 2).

Evidence of respiratory infection and severe congestion were disclosed by histology. The different fragments of lung have, roughly, a preserved architecture. It shows a suppurative exudate rich in neutrophils (Fig. 3a). The alveoli's lumen contains focally edematous lesion and is occupied by a dense cell population, consisting mainly of neutrophils and some macrophages ("leukocytic alveolitis") (Fig. 3b).

Other organs had no significant pathologic findings except for non-specific congestion.

Table 1 Data concerning heart weight and ventricular wall thicknesses in our case comparing with lower-upper 95% confidence limits (tables from Scholz et al, 1988.¹⁹

Organes	Our case	Lower-upper 95% confidence limits
Heart weight (g)	58 g	[12 g–33 g]
Left ventricular wall thicknesses (cm)	0.82 cm	[0.21 cm–0.84 cm]
Right ventricular wall thicknesses (cm)	0.43 cm	[0.06 cm–0.26 cm]
Ventricular wall thicknesses (cm)	0.92 cm	[0.22 cm–0.93 cm]

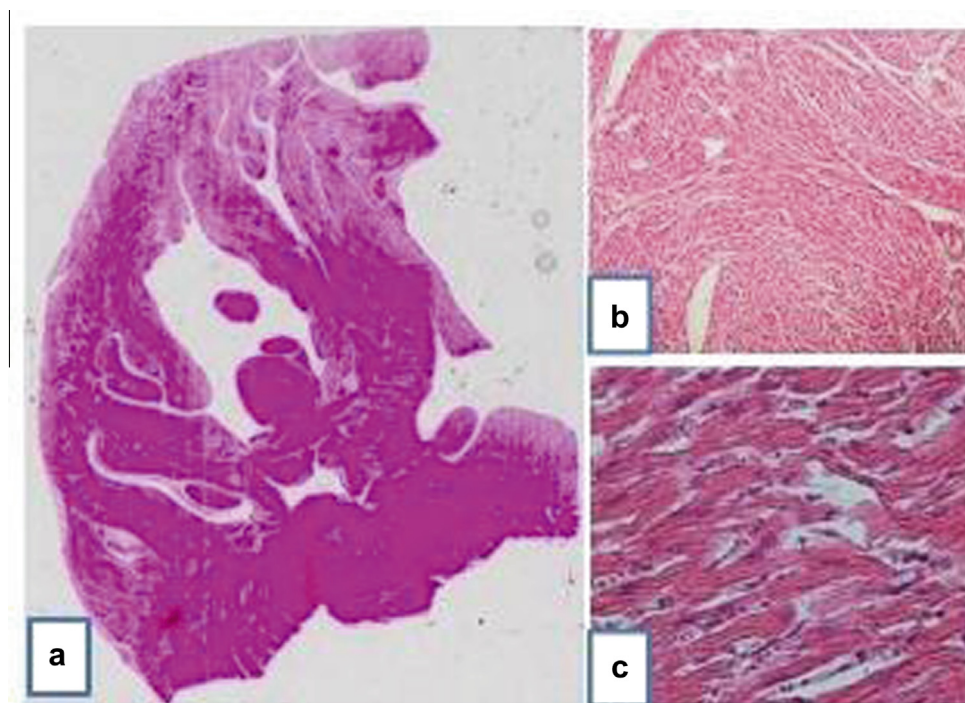


Figure 2 (a) Mounted slides: two chambers (arrow) separated by myocardial bundles simulating a septum (*). (b): HE staining $\times 200$: the septum separating the two chambers is composed by whorled myocardial fascicles. (c): HE staining $\times 400$: the myocardial cells have a hypertrophic nuclei.

To eliminate a toxic death, full toxicological examinations was performed producing only negative results.

Based on the autopsy findings, death was attributed to a congenital heart disease: isolated double-chambered right ventricle (DCRV). The manner of death was stated as natural.

3. Discussion

DCRV is a rare form of congenital heart disease in which the right ventricle is divided into a proximal high-pressure inlet portion and a distal low-pressure outlet portion by an anomalous muscle bundle that protrudes from the right ventricular free wall to the interventricular septum.¹⁵ The lesion makes up approximately 0.5–2% of CHD.⁴ Initially this entity was described in 1858 by Peacock.⁵

It can be caused by the presence of anomalous muscle tissue, hypertrophy of the endogenous trabecular bands, or an aberrant moderator band; all of which will typically result in progressive obstruction of the outflow tract.^{5–18}

Many classifications were proposed.⁶ A simple one proposed by Dr Folgers in 1996, who divided DCRV into two distinct types. The first type is characterized by an anomalous muscle bundle crossing the right ventricular cavity. In the second type there is no anomalous muscle bundle and the obstruction is caused by marked parietal and septal muscle hypertrophy.⁷

Often many cardiac anomalies are associated with DCRV. The most commonly associated cardiac anomaly is ventricular septal defect. It's associated with DCRV from 63% of cases in the study of Cil⁸ to 100% of the cases in the study of Choi.⁹

The other coexisting lesions include subaortic stenosis, pulmonary valve stenosis, double outlet right ventricle, tetralogy of Fallot, anomalous pulmonary venous drainage, complete or corrected transposition of the great arteries, pulmonary atresia with intact ventricular septum, and Ebstein anomaly.^{10,11} DCRV is exceptionally rare as an isolated anomaly.¹² Cil⁸ reported in his study that of 52 patients, five had isolated double chambered right ventricle.

No risk factors were associated with this anomaly. Some sporadic cases have been reported in patients with Down syndrome and Noonan syndrome.¹³

Obstruction to pulmonary blood flow usually progresses with hypertrophy of the muscle and further obliteration of the right ventricle cavity, this is why asymptomatic patients can become symptomatic afterward.¹⁴ The clinical exam may showed cyanosis, fatigue and decreased exercise tolerance, failure to thrive and growth retardation in children.^{9,14}

The most helpful complementary exams in patients with DCRV are transesophageal echocardiography, cardiac MRI and cardiac catheterization. They are required not only for confirming diagnosis but also for searching associated anomalies.¹⁶

The surgical repair is made by the resection of the obstructing muscle bundles. The long-term prognosis for patients after intracardiac repair is excellent.¹⁷

In our case the infant presented with, almost a week before death, shortness of breath and chest infection.

In our case the autopsy showed an isolated DCRV type 1. There are no reported cases of sudden unanticipated death in infancy due to isolated double-chambered right ventricle.

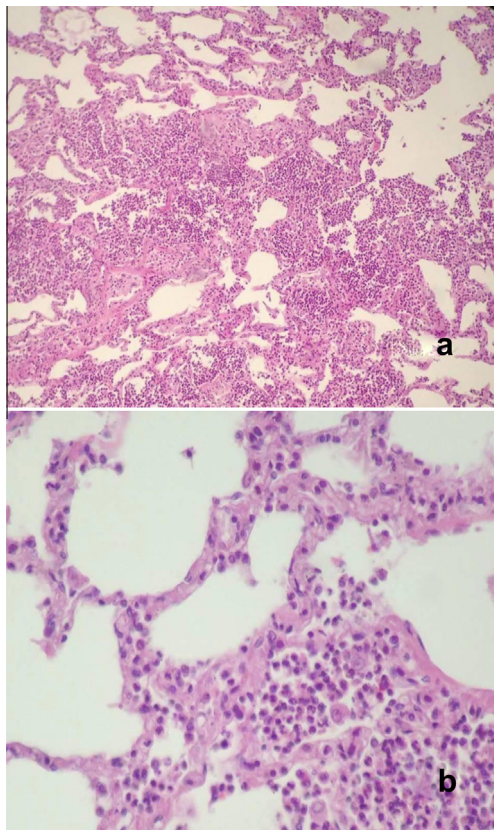


Figure 3 (a) Lung parenchyma H&E stain ($\times 40$): a preserved architecture of the lung with a suppurative exudate rich in neutrophils. (b): lung parenchyma H&E stain ($\times 100$): the alveoli's lumen contain focally edematous lesion and occupied by a dense cell population, consisting mainly of neutrophils and some macrophages.

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None.

Conflict of interest

None declared.

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